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Thrombocytopenia in Association With a Wandering Spleen

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An ectopic, so-called wandering spleen is an uncommon occurrence. We present the case of a young woman who presented with abdominal pain and was found to have an enlarged spleen, located in the lower abdomen and pelvis. The possibility of lymphoma was entertained because of concomitant findings of thrombocytopenia and a possible mesenteric mass. The mass was subsequently found at laparotomy to be the tail of the malpositioned pancreas, and the thrombocytopenia resolved with splenectomy. Review of the literature indicates that lymphoma is an uncommon finding in wandering spleens, that wandering spleens are enlarged in most cases, and that thrombocytopenia, while uncommon, can be seen, in particular when associated with torsion of an elongated splenic pedicle. © 1996 Wiley-Liss, Inc.

Key words: spleen, wandering spleen, ectopic spleen, pancreas, malposition, thrombocytopenia

INTRODUCTION

The spleen is usually fixed in its position in the left upper abdominal quadrant through the lienorenal and the gastrosplenic ligaments. The spleen gets additional support from contact with the phrenicocolic ligament. Malformation of these ligaments, their absence, or their laxity may lead to ptosis of the spleen and malposition in the lower abdomen or pelvis. Such a ptotic spleen is also referred to as a wandering, pelvic, or ectopic spleen. The rarity of this clinical entity, and the unfamiliarity of most clinicians with it, may lead to unfounded concerns about a malignant process, and may prompt unnecessary diagnostic procedures in a patient who has a wandering spleen.

CASE REPORT

A 30-year-old woman presented with a 1-month history of sudden onset left upper-quadrant abdominal pain without any associated symptoms. The pain spontaneously disappeared after several hours, but recurred several times in the next few weeks. Standing made the pain worse, while lying down resulted in partial relief. Food intake sometimes worsened the pain, but it was uninfluenced by bowel movements or urination. She had occasional vomiting when the pain was severe. Her gynecologist found no abdominal masses on physical examination, but pelvic examination revealed a firm, nontender, fixed mass in the right pelvis. Ultrasound confirmed a right adnexal mass, measuring 4.8 × 4.3 cm, of mixed density and containing calcifications. In addition, the spleen was reportedly enlarged.

The patient was scheduled for surgical removal of what was thought to be a benign ovarian teratoma. Preoperative blood count revealed a hemoglobin of 13.2 gm/dl, a white-cell count of 6,600/μl, and a platelet count of 104,000/μl. Because of the presence of splenomegaly and thrombocytopenia, abdominal and pelvic CT scans were obtained. The spleen was not in its usual position, but was located in the lower abdomen and in the pelvis (Fig. 1). The presence of a right adnexal mass with mixed attenuation was confirmed. An upper gastrointestinal and small-bowel follow-through barium study was normal except for slight displacement of small-bowel loops to the right of the midline.

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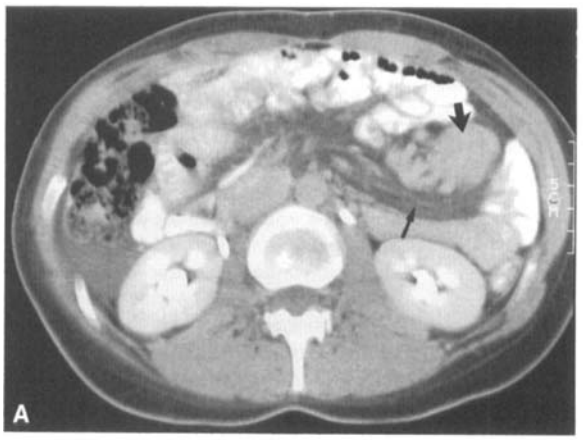
The patient's past medical history was significant for easy, but not excessive, bruising, but no other hemorrhagic problems. She had never been pregnant, and reportedly had a normal platelet count 3 years prior to presentation. Physical examination revealed no lymphadenopathy, hepatomegaly, or left upper-quadrant masses. There was mild tenderness in the periumbilical region, and a fullness without a definite mass was noted in the midline below the umbilicus. Review of the peripheral blood film confirmed mild thrombocytopenia, but was otherwise unremarkable. Prothrombin time was minimally prolonged at 12.7 sec (normal range, 10.3–12.5 sec). Activated partial prothrombin time was normal. Fibrinogen was 187 mg/dl (normal range, 146–410 mg/dl), and a D-dimer test was slightly elevated at 0.25–1.25 mg/dl (normal, <0.25 mg/dl). Factor V and VII activity levels were normal. Bone-marrow aspirate and biopsy were unremarkable, and a direct platelet antibody test was negative.

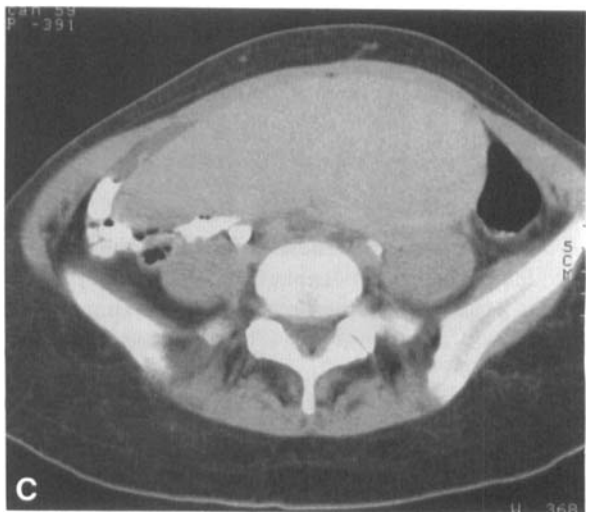
At laparotomy the spleen was located in the midabdomen, oriented in a transverse direction on top of the intestines. The spleen was free-floating, except for attachment to a greatly enlarged and elongated splenic pedicle. There was no torsion of the pedicle. The spleen was enlarged, but without acute congestion. The tail of the pancreas was displaced from its usual transverse location and was oriented up-and-down, lying intraperitoneally and across the intestines, running within the splenic pedicle towards the hilum of the spleen. The liver was normal in appearance and location. There were small paraaortic lymph nodes. The spleen and a right ovarian teratoma were removed, and several intraabdominal lymph nodes were sampled. The spleen weighed 330 g and contained several small subcapsular simple cysts, measuring up to 1 cm. Histological sections showed sinusoidal congestion, but were otherwise unremarkable. Histology of all sampled lymph nodes was also unremarkable. There was no evidence of lymphoma.

One week after splenectomy the patient's platelet count was 855,000/l. On follow-up 15 months after surgery, the patient was asymptomatic and doing well. She had not had any further occurrences of abdominal pain. Her platelet count was 358,000/μl.

DISCUSSION

A wandering spleen is rare, with only 6 cases reported among 3,873 collected patients who underwent splenectomy for various splenic problems [1–3]. In total, fewer than 500 cases of wandering spleen have been reported in the literature. Abell [4] in a literature review in 1933 identified 97 cases of wandering spleen with torsion of the pedicle; he did not include any cases in which torsion of the pedicle was not specifically mentioned as a causative factor in the production of symptoms and pathology. Lamesch and Lamesch [5] reviewed the pediatric literature from 1896–1990 and identified 74 cases of malposition of the spleen in children under age 14 years. Rodkey and Macknin [6] also reviewed the English pediatric literature and identified 51 cases of wandering spleen in children up to age 10 years between 1900–1991. Lastly, in a review of the English literature from 1960–1992, Dawson





and Roberts [7] identified 148 cases, in both children and adults, of wandering spleen.

Wandering spleen is diagnosed relatively more commonly in children than in adults, but patients can come to medical attention at any age. Dawson and Roberts [7] found that under age 10 years, the sex distribution was even, whereas over age 10, females outnumbered males 7:1. Abell [4] in his 97 cases of wandering spleen observed that women were affected 13 times more frequently than men. The higher incidence of wandering spleen in women of childbearing age suggests that pregnancy, through hormonal effects, may lead to laxity of the supporting splenic ligaments and to ligamentous lengthening.

The splenic pedicle is formed by the splenorenal and gastrosplenic ligaments and contains the splenic artery and vein, and the tail of the pancreas. In cases of wandering spleen, a lengthened splenic pedicle is always found. Congenital anomalies of various splenic ligaments have been proposed as etiologic factors. Malformation or absence of the gastrosplenic ligament is noted in many cases [8]. Numerous cases in which the splenorenal ligament is absent have also been reported [9], as was seen in our patient. This absence appears to be due to incomplete developmental fusion of the mesogastrium. It has been suggested that flacidity of the abdominal wall may also play a role in the development of a wandering spleen, and that this factor may explain the higher incidence in women of childbearing age [10]. The elongated pedicle predisposes the spleen to torsion, either acute or chronically recurrent, with possible infarction. In many reports of wandering spleen, similar to the case presented in this paper, the pancreas has been reported to lie in the splenic pedicle and can, in cases of splenic torsion, actually be involved in the twisting process [8].

Most pelvic spleens that come to medical attention are enlarged. Abell [4] noted that out of 53 patients for whom spleen weight was given, 45 had a spleen weight >500 g, i.e., 85%. Buehner and Baker [13] noted in their review of 140 cases of wandering spleen that most spleens were described as enlarged or congested, and that torsion of the pedicle was noted in most reports. Progressive splenomegaly of any cause may lead to enhanced splenic mobility because of the weight of the organ put onto the supporting ligaments. This does not lead to a significant elongation of the splenic pedicle, however, which would lead to an ectopic spleen. Splenomegaly generally appears to not be a predisposing factor to a wandering spleen, but a consequence thereof. This is evidenced by the fact that ptosis of the spleen is not often found in those parts of the world, where splenomegaly in its different forms (as in malaria) is endemic. Lamesch and Lamesch [5] found that in 74 pediatric cases of wandering spleen, the reason for enlargement was always the resut of torsion with subsequent splenic congestion, and never splenic disease. Torsion or compression of the splenic pedicle with impairment of venous outflow and resultant splenic congestion may lead to splenic enlargement.

Thrombocytopenia in patients with wandering spleen has rarely been reported. Most reports do not mention the patients' platelet counts. When reported, platelet counts are typically either normal or elevated. A total of 6 patients with thrombocytopenia and wandering spleen has been described in the literature (Table I). With the exception of our patient, all patients manifested torsion of the splenic pedicle. A few cases of leukopenia together with anemia have also been reported, possibly suggesting hypersplenism. Much more commonly, however, leukocytosis is found.

With ectopic pelvic spleen being a rare entity, pathological involvement of a pelvic spleen is an even rarer occurrence. There are only four reports in the English literature of involvement of a wandering spleen with malignancy (Table II). All four cases had a systemic malignant lymphomatous disease, and all four cases were middle-aged to elderly patients, as opposed to our relatively young patient.

Patients with wandering spleens may be asymptomatic or may have symptoms such as pelvic discomfort, abdominal or pelvic pain, disturbance of menstruation, dysuria, or urinary incontinence, or symptoms of gastrointestinal obstruction. Chronic and acute torsion of the spleen with infarction may lead to abdominal catastrophe. Splenectomy, splenopexy, and noninvasive, observant management have all been employed as treatment modalities for wandering spleen. In asymptomatic patients, splenopexy or splenectomy appear indicated to prevent torsion, infarction, and abdominal catastrophe. In patients presenting with acute abdominal findings, but in whom there is no evidence of thrombosis or infarction, detorsion and fixation should be considered, particularly in young patients most at risk for infection and postsplenectomy sepsis. All acute cases, in which splenic infarction has occurred, require splenectomy.

In our patient, the presence of a markedly elongated

Fig. 1. Images of abdominal and pelvic CT scans. A: Level of midabdomen shows a soft tissue mass at left (thick arrow), initially interpreted as a possible intussuception or a mass arising within the mesentery or small bowel. Laparotomy showed this mass to be the tail of the malpositioned pancreas. Within left upper abdominal quadrant, note abnormal appearance of the small bowel and mesentery, with stranding within the mesentery (thin arrow). B: Level of lower abdomen below kidneys. C: Level of midpelvis. B and C show malpositioned, markedly enlarged spleen. Soft tissue mass seen in A extends caudally to the hilum of the spleen (thick arrow in B), and correlates with the surgical finding of the tail of the pancreas, which lay intraabdominally and accompanied the splenic pedicle to the splenic hilum. CT scans show no abdominal or pelvic lymphadenopathy.

TABLE I. Reports in the Literature of Patients With Wandering Spleen Associated With Thrombocytopenia*

Reference	Patient age/sex	Platelet count (/μl)	HBg (g/dl)	WBC (/μl)	Operative findings	Spleen size/weight	Treatment
Weinreb et al. 1974 [11]	46/F	60,000	12.3	2,700	Torsion × 2	Enlarged/763 g	Splenectomy
Vermylen et al., 1983 [12]	13/F	100,000	13.3	6,300	Torsion	Enlarged/NR	Splenectomy
	12/F	110,000	"Normal"	8,500	Torsion	Enlarged/NR	Splenectomy
Riebel et al., 1985 [10]	14/F	64,000	NR	NR	Torsion × 7	Enlarged/320 g	Partial resection and splenopexy
Buehner and Baker, 1992 [13]	21/F	"Signif. decreased"	"Signif. decreased"	"Signif. decreased"	Torsion × 1	Enlarged/624 g	Splenectomy
This report	30/F	104,000	13.2	6,600	No torsion	Enlarged/330 g	Splenectomy

*NR, not reported; F, female; Signif., significantly.

TABLE II. Malignancy Involving Wandering Spleen: Literature Review of Cases*

Reference	Patient age/sex	Diagnosis	Comments
Waldman and Suissa, 1978 [14]	65/F	Lymphosarcoma	Extensive lymphosarcoma, involving, among other organs, an ectopic pelvic spleen
Barloon and Lu, 1984 [15]	85/F	Lymphoma	Splenic hilar lymph nodes and gallbladder also involved
Kinori and Rifkin, 1988 [16]	58/F	Lymphoma	Liver and abdominal lymph nodes also involved
Dautenhahn et al., 1989 [17]	64/F	Lymphoma	Liver and multiple lymph nodes also involved

*F, female.

splenic pedicle, the absence of the splenorenal ligament, and the intraperitoneal malpositioned tail of the pancreas indicate a congenital maldevelopment. The mesenteric mass, interpreted on CT scan as a possible neoplastic process or an intussusception, revealed itself during laparotomy as the tail of the pancreas. Enlargement of the spleen may have been due to chronic congestion secondary to positional dependence or secondary to intermittent splenic pedicle torsion or positional splenic vein compression with venous outflow obstruction. This would explain the intermittent nature of the patient's abdominal pain. The thrombocytopenia most likely was due to hypersplenism.

Knowledge of the clinical and laboratory associations that can be seen with a wandering spleen can help in formulating a diagnostic plan for the individual patient. First, although most of these patients do have an enlarged spleen, lymphoma is an uncommon finding, especially in younger patients. Second, the pancreatic tail can be malpositioned, extending into the elongated splenic pedicle, which can be potentially confused with an intraabdominal mass. Finally, although thrombocytopenia appears to be an uncommon finding, it can be seen with wandering spleen, although most frequently in association with torsion of the pedicle.

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